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A CASE OF COLLOID DISEASE OF THE BLOOD VESSELS OF THE SPINAL CORD.[1]

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The following case is unique in my experience and I have been unable to find any other like it in the literature. As the title indicates, it is one of focal lesion of the cord due to colloid disease of its blood vessels. The history is briefly as follows:

M. H., age forty-three, white; occupation, domestic.

Family history—Father and mother both killed by accident; otherwise negative.

Previous history—Has had the various diseases of childhood. Menstruation began at eighteen years, then ceased for a year, returned and occurred regularly every two weeks for two years. Subsequently it continued regularly every third week up to the present time. Suffered from displacement two years ago from which she recovered under treatment. Four years ago she had a slight attack of influenza, characterized by severe pains in neck, back and chest. About this time also was attacked by diarrhea which patient attributed to exposure to cold and wet. She characterized the diarrhea as uncontrollable. She continued to be up and around, however, and attended to her work. At the end of a year the diarrhea ceased and she began to slowly lose power in both hands, especially in the right. She was able, however, still to walk without difficulty.

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[1] Read at the annual meeting of the American Neurological Association, June 5, 6 and 7, 1902.

About a year later she began to have pains in the back, muscular rheumatism and stiffness of the joints. Her condition at the time of admission was as follows:

She stood with difficulty apparently because of stiffness in the joints of the legs and feet. There was stiffness of all of the joints, particularly of the smaller ones. There was a slight contraction of the middle and ring fingers of the right hand. Ankle-clonus was not noted, nor was there noted any sensory loss. The general physical examination was negative, save that both sounds of the heart appeared to be slightly roughened and there was a partial opacity of the left cornea, the result of scarlet fever.

Her condition presented from time to time little change. Examined a year later, it was found that the muscles of the shoulder had become distinctly atrophic, the fingers were contracted, the legs spastic, there were present exaggerated knee-jerks and ankle-clonus. Ankle-clonus was more marked on the right side. There was also a slight scoliosis. The gait was spastic, there was no involvement of the bladder or bowels and no trophic lesions.

There was now noted a marked anesthesia on the outer aspect of the left arm and left forearm. The thermal sense was also involved in this area; the patient made repeated mistakes when tested for hot and cold. No sensory losses were noted elsewhere. An examination of the eye at this time revealed the pupillary reactions to be normal. No view of the fundus of the left eye could be obtained because of opacity of the cornea. In the right eye the fundus was normal. An examination of the urine was negative.

Little by little the spasticity and contracture of the arms and legs became more and more pronounced, until in March, 1898, she became unable to walk. The loss of the power to walk was exceedingly gradual. At various times she suffered from pain in the back and from severe pain in the arms and legs. The bowels also became markedly constipated and in addition there was difficulty in micturition; the patient was obliged to wait for quite a while before the bladder could be emptied and at times it became necessary to use the catheter. No trouble was noted with speech or deglutition. She complained of numbness of the right hand and of the right leg.

Examined in detail, July 29, 1898, she revealed the following: The patient is bed-ridden. She cannot move the right leg at all. She can flex and extend the toes of the right foot a little. She can barely lift the left leg from the bed, but can flex and extend the left leg feebly. She can abduct and adduct the left thigh a little. She can move the left foot and toes fairly well.

She cannot move the right arm at the shoulder at all, but can flex and extend the right arm feebly. There is almost no grip. She can move the left arm at the shoulder quite well and can flex and extend the left forearm quite well; also pronate and supinate the left forearm. There is no palsy of the face. The tongue is protruded straight and is not wasted. There is marked wasting of the intrinsic muscles of both hands, arms and forearms. There is no wasting of the legs or thighs. Insteps of both feet are very high. Patellar jerk is very marked on both sides. Patellar clonus is present on both sides. Ankle-clonus is marked on the right side, slight on the left. Plantar jerk present on both sides.

The sensory losses were now found to be more marked:—

Tactile sensation is abolished over the whole left upper extremity. There is no sensory loss over the right upper extremity except some impairment of sensation over the dorsal surface of the hand and fingers and a doubtful impairment over the palmar surfaces of the fingers. There is complete tactile loss over the left side of the chest, covering an area bounded above by the clavicle, below by the sixth rib, laterally by the middle line of the body. In the lower extremities, sensation is intact. An area of tactile anesthesia is noted, about two inches in diameter, extending downward and inward from the right nipple. There is tactile loss also on the left side of the neck; the face and ear, however, are not involved. On the upper portion of the back, on the left side, there is a complete tactile loss over an area extending from the junction of the neck and trunk above, downward to about two inches below the angle of the scapula and from the spine to the anesthetic area on the left side of the chest with which it merges.

The *pain* sense also reveals marked anomalies. There is complete loss to pain over the whole of the left upper arm, posterior surface of the left forearm and on the back of the left hand in an area corresponding to the first, second and third metacarpal bones and their respective fingers. There is a partial loss of the pain sense over the anterior surface of the left forearm, over the dorsal and palmar aspects of the hand in the region corresponding to the fourth and fifth metacarpal bones and over their respective fingers. There is complete loss of the pain sense over the left chest covering an area bounded above by the first interspace, below by the fifth interspace and internally by the middle line of the body. The pain sense was intact over the right side of the chest, the right upper extremity and both lower extremities. On the upper portion of the back, on the left side, there is complete loss of the pain sense over an area extending from the spine of the scapula down to the level of its angle; it does not extend entirely to the spinal column and laterally merges with the area of pain loss on the left side of the trunk. Partial loss of pain sense extends from the posterior border of the scapula over to the spine and downward as far as the limits of the area for tactile loss.

The *thermal* sense is abolished entirely over the left arm and posterior part of forearms, also on dorsal aspect of hand over first, second and third metacarpal bones and their respective fingers. It is almost entirely abolished over the area for loss of pain on the left chest; here hot and cold are confused, more usually cold being mistaken for hot. Over the anterior surface of the left forearm, there is a partial loss and confusion. Over the fourth and fifth metacarpal bones and their respective fingers, of the left hand, both on the dorsal and palmar aspect, there is some impairment, but hot and cold are usually called correctly. Over the posterior portion of the trunk, the thermal loss practically follows that of pain, but in addition includes the lower half of the ear and margin of the face about the angle of the jaw. It also extends toward the spine to a line about half way between the border of the scapula and the spine. From this line over to the spine, hot and cold can be distinguished. From one-half to one inch below the angle of the scapula down to the limit of the area of tactile loss, there is a partial loss of thermal sensations. This corresponds to the area for partial painful loss. The thermal sense is intact over the whole of the left thigh and leg, also over the right thigh. From knee down, over the whole of the right leg, there is an impairment which seems to vary at different parts, but is most pronounced over the internal malleolus.

November 9, 1898. Patient's condition practically unchanged. Complains now of tenderness in the ankles in addition to weakness. States now that she has always suffered considerably from pain in the joints and that this pain was made worse on motion. Mental condition somewhat obtunded so that she may be in error with regard to the past history of pain.

Ophthalmoscopic examination at this time again gave a negative result.

November 25, 1898. Patient now complains of a sensation of constriction about the waist. There is also diffuse tenderness to pressure over the calves, thighs and both arms. This tenderness is quite pronounced.

On November 30, 1898, the patient attempted to get out of bed. She fell, striking her head, causing a wound of the scalp half an inch in length. It was dressed antiseptically and subsequently healed promptly.

From this time on her condition changed gradually and almost imperceptibly.

She was again carefully examined on February 28, 1900. The symptoms had been very slowly progressive and but little change had taken place since the previous examinations. The following note was made:

Muscular wasting is now somewhat more pronounced. She cannot move the right arm at the shoulder. She can move it to a slight extent and flex it at the elbow. She can neither flex nor extend the fingers which are held in a position of semiflexion. The muscles of the arm, forearm and hand are all somewhat atrophied. The biceps jerk is increased and the muscle jerks of the forearm are increased. Sensation is quite well preserved in the right arm. She cannot move the left arm at the shoulder. She can flex and extend the left elbow, flex and extend the left fingers, but all the movements are weak. There is slight wasting of the muscles of the left hand and left forearm. The left biceps jerk is absent; the muscle jerk is slight. There is anesthesia of the left arm and left chest, the areas corresponding closely with those determined at previous examinations.

Both legs are edematous. Left leg can be slightly extended at knee, ankle and toes. Right leg can be flexed at knee very slightly, almost no movement in hip of left side, none in right hip.

Sensation to touch preserved in both legs and there is some hyperesthesia. There is no wasting of the legs. She localizes touch well in legs and feet. No fibrillary twitchings. She confuses heat and cold on both legs, sometimes answering correctly; at times wrongly. Edema of the feet.

Both knee-jerks are exaggerated. On the left side distinct ankle-clonus, also on right. Stroking either sole causes flexion of toes. No extension. Achilles jerk preserved on either side, but slight. Striking the sole causes flexion of entire foot. On the abdomen at right side, sensation to touch normal; on the left side, normal up to line of ribs. Heat and cold are felt perfectly well on abdomen. Marked constipation. Good control of the bladder. Occasionally dribbling.

Abdomen shows a freely movable irregular tumor, probably feces. There is no deformity of spine.

May 3, 1900. Patient is growing weaker. Diuretics have lessened the edema of lower extremities. Patient is very restless at night.

October 1, 1900. Patient is in very poor condition this morning. For past few days has had severe diarrhea with some blood in the stools; also considerable pain and bearing down; is very weak.

October 2, 1900. Has seemed much worse today. Pulse cannot be felt at the wrist and patient bathed in a cold sweat, with sub-normal temperature. During greater part of day has been unable to swallow any medicine or nourishment.

She died October 3, 1900.

The urine had been repeatedly examined during life and always with a negative result except with an occasional low specific gravity. Examination of the sputum had also yielded a negative result.

The body was removed from the hospital before an autopsy could be held. The resident physician, Dr. E. T. Robinson, followed the body to a distant part of the city and under discouraging circumstances succeeded in securing the spinal cord. Unfortunately no general post-mortem could be held. Microscopical examination revealed the following changes in the cord.

Sections from the upper part of the cervical swelling. The central portion of the spinal cord is much disintegrated so that the tissue in some parts has entirely disappeared; in other areas the tissue is very loose. This area of softening extends into the lateral column of each side, but more into the left than into the right lateral column. The crossed pyramidal and direct cerebellar tracts are degenerated. A glassy homogeneous infiltration in patches is seen in the central region of the cord, in the posterior columns, and in the lateral columns, especially in the left lateral column. This infiltration in some areas has replaced the normal tissue of the cord; in others it has penetrated between the nerve fibers leaving them more or less widely separated from one another and embedded in a homogeneous glassy matrix. Some much swollen axis cylinders are seen here and there in this infiltration. The walls of many of the intramedullary blood vessels are much thickened and have undergone a hyaloid change and in some more than one lumen is found. Disseminated sclerotic areas are found in different parts of the transverse section and not always in association with thickened blood vessels. Round-cell infiltration is not very distinct either in the cord or pia, although here and there a little infiltration may be seen about a vessel. Many of the nerve cell-bodies of the anterior horns, stained by ammonium carmine, are atrophied and with few or no dendritic processes. The anterior and posterior spinal roots appear to be normal, although the posterior possibly may be slightly degenerated. This, however, is very doubtful. Sections of the spinal cord above the upper part of the cervical swelling were not obtained. Sections from the middle part of the medulla oblongata are normal, except that the anterior pyramids were destroyed when the spinal cord was removed at the necropsy.

Sections of the cervical region taken from lower levels than those described above, present a similar picture. The nerve cell-bodies of the anterior horns, especially of the left horn, are very distinctly degenerated. Here and there a slight round-cell infiltration, about an intramedullary blood vessel, is found. The homogeneous infiltration of the cord—very pronounced at this level also—stains a light chocolate color by the Weigert's hematoxylin method and pale pink by ammonium carmine.

Sections from about the eighth cervical region are very similar to those described from higher levels, although the alteration, except in the crossed pyramidal tracts, is less intense. Here and there a small recent hemorrhage is seen within the spinal cord. Sections from this level stained by the Marchi method do not show any recent degeneration.

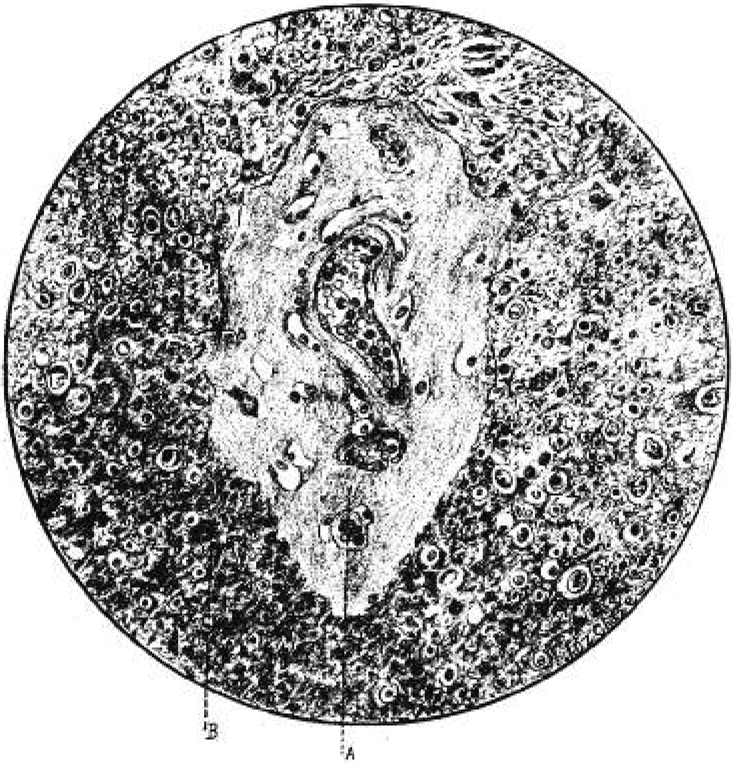

Fig. 1. Section from the upper portion of the cervical region. A, Blood vessel showing colloid-like degeneration of its walls. Three or four lumina are found within this vessel. B, Sclerotic portion surrounding degenerated blood vessel.

The degeneration of the crossed pyramidal tracts extends through the thoracic region into the lumbar.

The microscopical work was done by Dr. Wm. G. Spiller at the Pepper Clinical Laboratory.

It is to be regretted that the portion of the spinal cord above the cervical swelling was not obtained, but the findings are sufficient to explain the symptoms. Throughout the cervical swelling many of the blood vessels have undergone a hyaloid change, and a glassy infiltration, resembling that described by Obersteiner as colloid infiltration, has occurred, separating and destroying the normal tissue of the cord and causing softening of the cord and secondary degeneration.

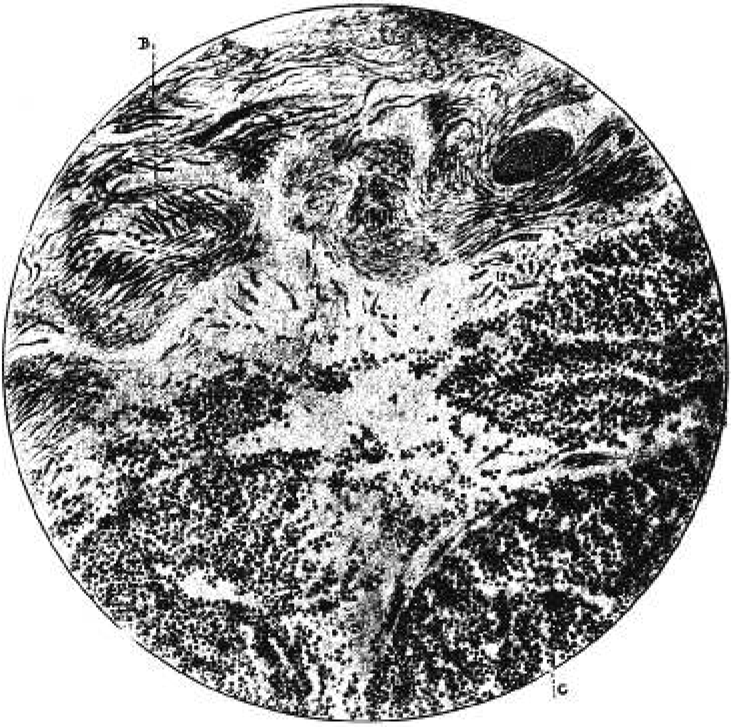

Fig. 2. Section from about the eighth cervical region. A, Homogeneous infiltration, staining light chocolate color by the Weigert hematoxylin method. B, Posterior horn and posterior commissure infiltrated by the homogeneous exudate. C, Posterior columns.

The only reference bearing upon this case, of which I have any knowledge, is that of Alzheimer[2], who describes two cases of colloid degeneration of the vessels of the brain. In one of these the symptoms were those of paresis, and Alzheimer regarded the case as one of paresis complicated by colloid disease of the blood vessels. In the second, the symptoms were those of brain tumor. The patient had during life choked disc, left-sided paralysis, convulsive attacks limited almost exclusively to the left side of the body, and headache. Numerous foci of softening and degeneration were found in the right hemisphere, in the right basal ganglia and in the right crus, together with colloid disease of the vessels.

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[2] Archiv f. Psychiatrie, Volume 30, page 18.

In the case here presented, a large focus of softening and infiltration was present in the upper cervical portion of the cord, besides there were areas of softening and infiltration present in various other portions as described. Colloid disease of the blood vessels of the central nervous system, presenting either diffuse or focal lesions, must be regarded as excessively rare and the case before us is unique. During life no definite diagnosis had been made. Early in the case syringomyelia and later on tumor of the cord seemed probable.